Malignant Fibrosarcoma of the Maxillary Sinus Masquerading as an Angiectatic Nasal Polyp: Case Report and Review of the Literature

Jordan Rawl, M.D.; Bailey LeConte B.S.; Suiman Qiu, M.D.; Mohamad Chaaban M.D., M.S.C.R, M.B.A
Department of Otolaryngology, Head and Neck Surgery
University of Texas Medical Branch, Galveston, Texas

Abstract

Objectives: To highlight the pathology and treatment strategies of myxofibrosarcoma by presenting a case report of this rare head and neck tumor.
Methods: A presentation of a case and a review of pertinent literature
Results: Myxofibrosarcomas are rare tumors of the head and neck. Our patient was initially diagnosed with a right rapidly growing angiectatic nasal polyp which on repeat, deep biopsy obtained in the operating room demonstrated low grade myxofibrosarcoma. In an attempt to spare his right eye, he was treated with neoadjuvant radiation. However, his tumor enlarged, prompting radical maxillectomy and final pathology demonstrated progression to a high-grade lesion.
Conclusions: This case highlights the need for deep biopsy in clinically suspicious lesions. Additionally, it prompts further inquiry into the role of neoadjuvant radiation in the treatment of these tumors.

Introduction

Despite being the most common soft tissue tumor in adults, myxofibrosarcoma is rare in the head and neck.[1,2] To the best of our knowledge, there have been only 22 cases reported in the head and neck with 5 cases reported in the maxillary sinus.[1,2,3] We present a case report of a 55 year-old man with a rapidly growing right maxillary sinus myxofibrosarcoma with a review of the literature.

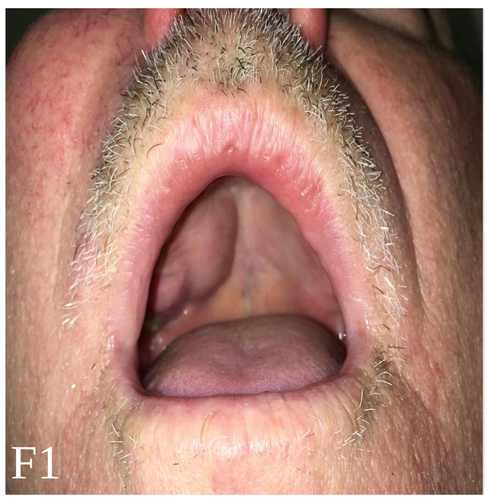
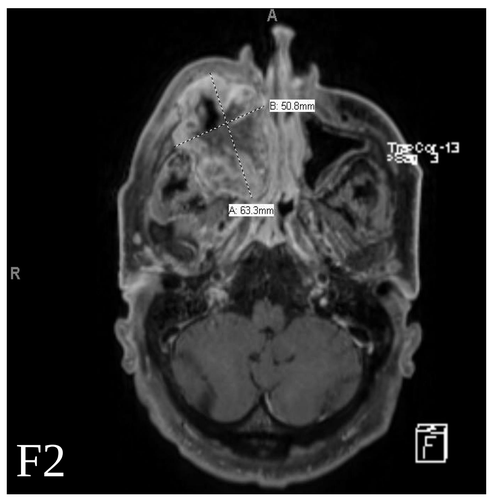


Figure 1 – Intraoral exam demonstrating bulging right hard palate. Also noted is hypervascularity and swelling of the right cheek.



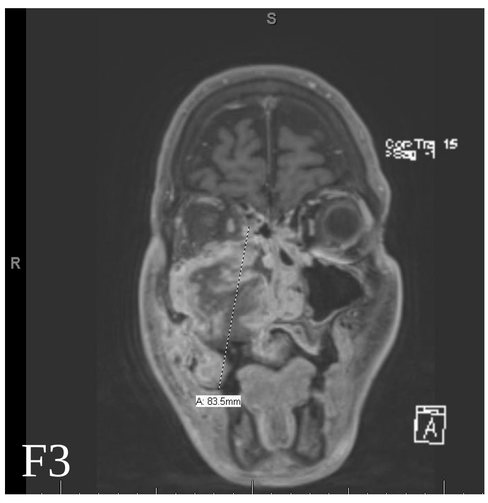


Figure 2 – T1 weighted MRI with contrast in the axial plane demonstrating cerebriform lesion eroding through the maxillary sinus wall anteriorly. Figure 3 – T1 weighted MRI with contrast in the coronal plane demonstrating lesion invading the right inferior orbit.

Case Report

Patient JM is a 55 year-old male cabinet-maker and former tobacco user with hypertension who presented to our clinic with a one year history of progressive right cheek swelling and a six month history of right nasal obstruction, epiphora and epistaxis. Head and neck exam revealed right cheek fullness, proptosis and a pale tan polypoid mass filling the right nasal cavity with bulging of the hard palate (**F1**). Initial contrast maxillofacial CT and MRI demonstrated a 5.5cm expansile right maxillary sinus mass extending into the nasal cavity, inferior orbit, eroding the anterior, superior, and posterior, walls of the maxillary sinus, and involving the posterior alveolar ridge and hard palate. Initial biopsies revealed necrotic, ulcerated tissue with granulation. The second set of biopsies, taken a week later, resulted in angioectatic nasal polyp. He was taken to the operating room and underwent endoscopic sinus surgery with deep biopsy of the mass. Permanent surgical pathology confirmed an atypical spindle cell neoplasm with myxoid changes (**F4**). Immunohistochemistry was negative for S-100, Desmin, MITF, HMB-45, and SMA. The final diagnosis was a low to intermediate grade myxofibrosarcoma. The patient subsequently underwent neoadjuvant radiation 50Gy in 25 fractions with a boost to 70Gy in an attempt to salvage the right eye. MRI and CT of the face and neck obtained one month after radiation treatment demonstrated interval increase in size of tumor measuring 6.3cm x 5.0cm x 8.3cm (**F2-F3**). Final treatment consisted of right total radical maxillectomy, orbital exenteration, selective neck dissection levels I-IV with reconstruction with a left anterolateral thigh free flap. Final pathology of the surgical specimen confirmed a cT4aN2cM0G2 high-grade myxofibrosarcoma (**F5**). The patient was seen three months post-operatively and was noted to be doing well with no recurrence.

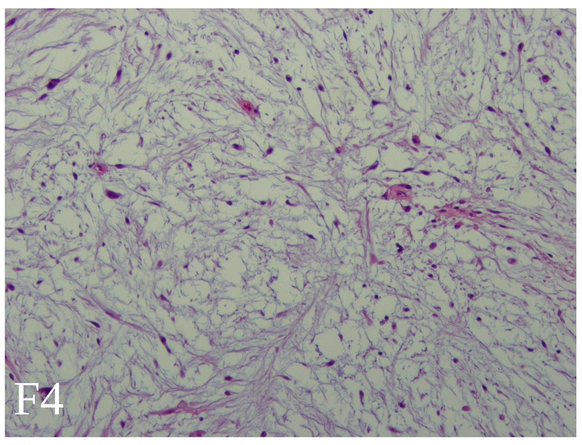


Figure 4 – Low-grade myxofibrosarcoma with hypocellular myxoid matrix with curvilinear vessels and atypical spindle cells (100x magnification).

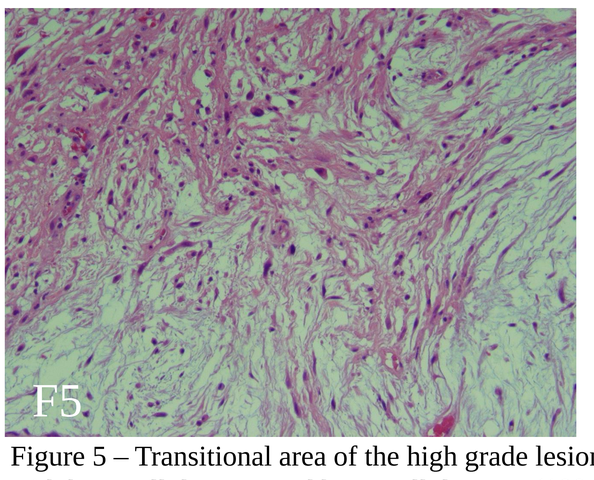


Figure 5 – Transitional area of the high grade lesion with hypocellular area and hypercellular area (100x magnification).

Discussion

Myxofibrosarcoma is a rare head and neck sarcoma. It usually presents in elderly patients, arising in the dermal or subcutaneous tissues of the limbs and trunk.[3] To our knowledge, no other case of maxillary myxofibrosarcoma has been misdiagnosed as an angiectatic nasal polyp. Myxofibrosarcomas classically consist of nodules of atypical spindle cells in a myxoid matrix divided by fibrous septa with curvilinear shaped capillaries and angiocentrically distributed lymphocytes and plasma cells.[2,5] Low grade tumors rarely metastasize but show a local recurrence rate as high as the higher grade tumors (50-60%).[2,3,4] Wide local excision with 3-5cm margins with adjuvant radiation therapy in margin positive cases are the mainstays of treatment. Chemotherapy has not been shown to be effective.[2,4] The overall 5-year and 10-year survival rates for patients with low-grade myxofibrosarcoma are 65% and 52% respectively.[2,4,6] Radiation as a single modality treatment has not been proven to be effective but its role as a neoadjuvant therapy has not been clarified.[7] Our patient was treated with neoadjuvant radiation and ultimately required radical surgery with final pathology demonstrating a high-grade lesion.

Conclusions

This case highlights the dire need for deep biopsy in clinically suspicious lesions. In our case two biopsies were performed in clinic demonstrating benign polyps. Additionally our case calls for further inquiry into the role of neoadjuvant radiation in the treatment of these tumors.

Contact: Jordan W Rawl, MD
University of Texas Medical Branch
Department of Otolaryngology – Head and Neck Surgery
301 University Blvd
Galveston, Texas 77551

References:
1. Nakahara, S et al. A case of myxofibrosarcoma of the maxilla with difficulty in preoperative diagnosis. Int J Clin Oncol (2012) 17:390-394
2. Qiubei et al. Myxofibrosarcoma of the sinus piriformis: case report and literature review. World Journal of Surgical Oncology 2012, 10:245
3. Dell'Aversana Orabona et al. Head and neck myxofibrosarcomas: a case report and review of the literature. Journal of Medical Case Reports 2014, 8: 468
4. Taghi AS, Ali A, Kuchai R, Saleh H. Rare sarcoma presented as sinusitis. BMJ Case Reports 2012; doi:10.1136/bcr-02-2012-5863
5. Norval EJG, Raubenheimer EJ. Myxofibrosarcoma Arising in the Maxillary Sinus: A case Report with a Review of the Ultrastructural Findings and Differential Diagnoses. J. Maxillofac. Oral Surg. (Oct-Dec 2011) 10 (4):334-339.
6. Fayda et al. The role of surgery and radiotherapy in treatment of soft tissue sarcomas of the head and neck: Review of 30 cases. Journal of Cranio-Maxillofacial Surgery (2009) 37, 42-48.
7. Fletcher CDM. Distinctive Soft Tissue Tumors of the Head and Neck. Mod Pathol 2002; 15 (3):324-330.